

Serial No. 10/826,112
Atty. Docket No.: P71641US0

Amendments to the Claims

Listing of Claims:

1. (Withdrawn) A method for treating a subject suffering from phenylketonuria and/or phenylalanemia, said method comprising the steps of:

obtaining a LNAA supplement in which the weight ratio of Leu to Val is greater than 2:1; and

enterally administering to the subject said LNAA supplement.

2. (Withdrawn) The method according to claim 1, wherein the LNAA supplement is substantially free from phenylalanine.

3. (Withdrawn) A method for treating a subject suffering from phenylketonuria and/or phenylalanemia, said method comprising the steps of:

obtaining a LNAA supplement in which the weight ratio of Leu to iLeu is greater than 3:1; and

enterally administering to the subject said LNAA supplement.

4. (Withdrawn) The method according to claim 3, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

5. (Withdrawn) The method according to claim 4, wherein the LNAA supplement is substantially free from phenylalanine.

6. (Withdrawn) A method for treating a subject suffering from phenylketonuria and/or phenylalanemia, said method comprising the steps of:

obtaining a LNAA supplement which comprises one or more LNAAs and which further comprises Lys; and

enterally administering to the subject said LNAA supplement.

7. (Withdrawn) The method according to claim 6, wherein the LNAA supplement comprises Leu.

8. (Withdrawn) The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1.

9. (Withdrawn) The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

10. (Withdrawn) The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

11. (Withdrawn) The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

12. (Withdrawn) The method according to claim 6, wherein the LNAA

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supplement comprises Leu; wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1; and wherein the weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

13. (Withdrawn) The method according to claim 12, wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

14. (Withdrawn) The method according to claim 12, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

15. (Withdrawn) The method according to claim 6, wherein the LNAA supplement comprises Leu; said weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1; and said weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

16. (Withdrawn) The method according to claim 6, wherein the LNAA supplement is substantially free from phenylalanine.

17. (Currently amended) A large neutral amino acid (LNAA) supplement comprising Leu and Val, ~~in which~~ wherein the weight ratio of Leu to Val is greater than 2:1.

18. (Previously presented) The LNAA supplement according to claim 17, wherein the LNAA supplement is substantially free from phenylalanine.

19. (Currently amended) A LNAA supplement comprising Leu and iLeu,

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~~in which~~ wherein the weight ratio of Leu to iLeu is greater than 3:1.

20. (Previously presented) The LNAA supplement according to claim 19, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

21. (Previously presented) The LNAA supplement according to claim 20, wherein the LNAA supplement is substantially free from phenylalanine.

22. (Currently amended) A LNAA supplement comprising one or more LNAAs and further comprising Lys in an amount between about 5 to 30 mg per 500 mg of LNAA supplement.

23. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement further comprises Leu.

24. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu and said weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1.

25. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu and said weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

26. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu, and said weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

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27. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu, and said weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

28. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu; said weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1; and said weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

29. (Previously presented) The LNAA supplement according to claim 28, wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

30. (Previously presented) The LNAA supplement according to claim 28, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

31. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu; said weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1; and said weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

32. (Previously presented) The LNAA supplement according to claim 22, wherein the LNAA supplement is substantially free from phenylalanine.

33. (Currently amended) The LNAA supplement according to claim 22, wherein the LNAA supplement comprises, per 500 mg of LNAA

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supplement:

| | | | | |
|------------|-----|-------------|--------------------------|----------------|
| from about | 100 | mg to about | 290 | mg of Tyr; |
| from about | 25 | mg to about | 75 | mg of Trp; |
| from about | 15 | mg to about | 50 | mg of Met; |
| from about | 15 | mg to about | 55 | mg of iLeu; |
| from about | 15 | mg to about | 50 | mg of Threo; |
| from about | 15 | mg to about | 55 | mg of Val; |
| from about | 15 | mg to about | 200 | mg of Leu; |
| from about | 10 | mg to about | 30 | mg of His; and |
| from about | 5 | mg to about | 200 <u>30</u> | mg of Lys. |

34. (Cancelled).

35. (Previously presented) The LNAA supplement according to claim 33, wherein the LNAA supplement is substantially free from arginine.

36. (Previously presented) The LNAA supplement according to claim 33, wherein the LNAA supplement is substantially free from phenylalanine.

37. (Original) A LNAA supplement comprising, per 600 mg of LNAA supplement:

| | | | | |
|------------|-----|-------------|-----|--------------|
| from about | 100 | mg to about | 290 | mg of Tyr; |
| from about | 30 | mg to about | 90 | mg of Trp; |
| from about | 25 | mg to about | 75 | mg of Met; |
| from about | 15 | mg to about | 45 | mg of iLeu; |
| from about | 15 | mg to about | 50 | mg of Threo; |

from about 15 mg to about 50 mg of Val;
from about 40 mg to about 200 mg of Leu;
from about 15 mg to about 45 mg of His; and
from about 15 mg to about 50 mg of Arg.

38. (Previously presented) The LNAA supplement according to claim 37, wherein the LNAA supplement further comprises Lys.

39. (Previously presented) The LNAA supplement according to claim 37, wherein the LNAA supplement further comprises, per 600 mg of LNAA supplement, about 5 mg to about 200 mg of Lys.

40. (Previously presented) The LNAA supplement according to claim 37, wherein the LNAA supplement is substantially free from phenylalanine.

41. (Withdrawn) A method for treating a subject suffering from a metabolic disorder involving the metabolism of a first amino acid X, said method comprising the steps of:

obtaining a composition which is substantially free from said first amino acid X, and said composition comprises a second amino acid Y that competes with amino acid X at a gastrointestinal tract transporter; and

enterally administering to the subject said composition.

42. (Withdrawn) The method according to claim 41, wherein the metabolic disorder is not phenylketonuria and/or phenylalanemia.

43. (Withdrawn) The method according to claim 42, wherein the

gastrointestinal tract transporter is a Caco-2 cell transporter.

44. (Withdrawn) The method according to claim 41, wherein the gastrointestinal tract transporter is a Caco-2 cell transporter.

45. (Withdrawn) The method according to claim 44, wherein the metabolic disorder is tyrosinemia; and the first amino acid X is tyrosine and the second amino acid Y is selected from the group consisting of Phe, Leu, Trp, Lys, His, and combinations thereof.

46. (Withdrawn) The method according to claim 44; wherein the metabolic disorder is tyrosinemia; the first amino acid X is selected from the group consisting of phenylalanine, tyrosine, and combinations thereof; and wherein the second amino acid Y is selected from the group consisting of Leu, Trp, Lys, His, and combinations thereof.

47. (Withdrawn) The method according to claim 44, wherein the metabolic disorder is alkaptonuria; the first amino acid X is selected from the group consisting of phenylalanine, tyrosine, and combinations thereof; and wherein the second amino acid Y is selected from the group consisting of Leu, Trp, Lys, His, and combinations thereof.

48. (Withdrawn) The method according to claim 44, wherein the metabolic disorder is homocystinuria; the first amino acid X is methionine; and the second amino acid Y is an amino acid that competes with methionine at a gastrointestinal tract transporter.

49. (Withdrawn) The method according to claim 44, wherein the

metabolic disorder affects metabolism of a branched amino acid selected from the group consisting of leucine, isoleucine, valine, and combinations thereof; said first amino acid X is selected from the group consisting of leucine, isoleucine, valine, and combinations thereof; and said second amino acid Y is an amino acid that competes with the first amino acid X at a gastrointestinal tract transporter.

50. (Withdrawn) The method according to claim 49, wherein the metabolic disorder is selected from the group consisting of maple syrup urine disease, isovaleric acidemia, methylmalonic acidemia, and propionic acidemia.

51. (Withdrawn) The method according to claim 41, wherein said method further comprises the step of restricting the subject's dietary intake of the first amino acid X.

52. (Withdrawn) The method according to claim 41, wherein said method further comprises the step of not restricting the subject's dietary intake of the first amino acid X.

53. (Withdrawn) The method according to claim 52, wherein said enteral administration is carried out substantially at mealtime.

54. (Withdrawn) The method according to claim 41, wherein said enteral administration is carried out substantially at mealtime.

55. (Withdrawn) The method according to claim 41, wherein said enteral administration is carried out orally.

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56. (New) A LNAA supplement comprising Leu and Val, wherein the weight ratio of Leu to Val is greater than 2:1, and wherein the LNAA supplement comprises, per 500 mg of LNAA supplement:

| | | | | |
|------------|-----|-------------|-----|----------------|
| from about | 100 | mg to about | 290 | mg of Tyr; |
| from about | 25 | mg to about | 75 | mg of Trp; |
| from about | 15 | mg to about | 50 | mg of Met; |
| from about | 15 | mg to about | 55 | mg of iLeu; |
| from about | 15 | mg to about | 50 | mg of Threo; |
| from about | 15 | mg to about | 55 | mg of Val; |
| from about | 15 | mg to about | 200 | mg of Leu; |
| from about | 10 | mg to about | 30 | mg of His; and |
| from about | 5 | mg to about | 200 | mg of Lys. |